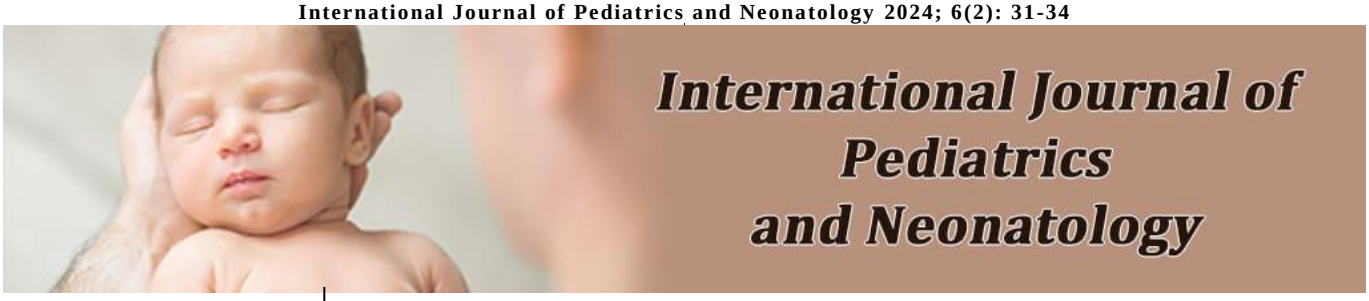

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Giant coronary artery aneurysms in Kawasaki disease: A case report

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Abstract
Kawasaki disease is one of the most common vasculitis of childhood, typically self-limited but may progress to the development of coronary artery aneurysms, which occurs in about 25% of untreated cases, and 3-5% of cases when adequate treatment is performed.
We report a case of Kawasaki disease that, despite adequate treatment in the first 10 days, evolved to the development of giant aneurysms of the coronary arteries, which persist after a year of follow-up.

Keywords: Kawasaki disease, small vessel vasculitis, acquired heart disease, coronary aneurism

Introduction
Kawasaki disease is a vasculitis syndrome primarily affecting children under the age of five. The exact etiology of Kawasaki disease remains elusive, but it is believed to involve immune dysregulation triggered by various infectious agents or environmental factors in genetically susceptible individuals.
The hallmark clinical features of Kawasaki disease include prolonged fever, mucocutaneous manifestations such as conjunctivitis, changes in the oral mucosa (strawberry tongue, oral mucosal erythema), extremity changes (swollen hands and feet), and cervical lymphadenopathy. These symptoms typically occur in a characteristic sequence but may not all manifest simultaneously.
If left untreated, Kawasaki disease can lead to serious complications, most notably coronary artery abnormalities (such as coronary artery dilatation and aneurysms), and myocarditis. Therefore, early recognition and prompt initiation of treatment are essential to prevent these potentially life-threatening complications.
The diagnosis of Kawasaki disease is primarily clinical, based on the presence of characteristic signs and symptoms. Laboratory tests may reveal nonspecific markers of inflammation. Echocardiography is essential for evaluating cardiac involvement and monitoring for coronary artery abnormalities.
Treatment typically involves intravenous immunoglobulin (IVIG) administration and aspirin therapy to reduce inflammation and decrease the risk of coronary artery complications. Additional therapies may be considered for refractory cases or those with severe coronary artery involvement.

Case Report
A previously healthy 3-year-old male child was evaluated at the emergency department due to 6 days of high fever, odynophagia and asthenia. On physical examination, cheilitis, raspberry tongue, bilateral non-exudative conjunctivitis, scattered rash on the trunk, perineal rash and desquamation, edema and erythema of the palms of the hands and bilateral cervical adenopathies were observed. Hypotension and gallop rhythm on cardiac auscultation were also detected.
A complementary analytical study revealed anemia, leukocytosis with neutrophilia and thrombocytopenia; hypoalbuminemia and hypoproteinemia; elevation of transaminases, gamma-glutamyltransferase and alkaline phosphatase; hyperbilirubinemia; hypertriglyceridemia; hyponatremia; elevation of acute phase markers including C-reactive

protein, procalcitonin and ferritin. It was also detected an elevation of troponin I and B-type natriuretic peptide; alteration of the coagulation study with reduced activated partial thromboplastin clotting time, prolonged prothrombin time and elevation of fibrinogen and d-dimers; as well as proteinuria and sterile pyuria. Most common etiological agents of infection were excluded.

Sinus tachycardia was found on the electrocardiogram, with no other relevant alterations.

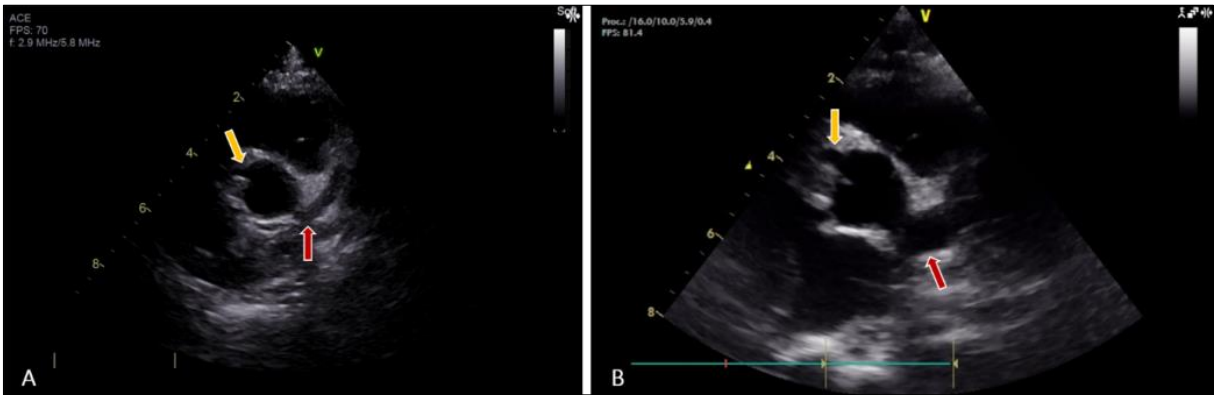
The transthoracic echocardiogram described structurally normal heart, mild depression of ventricular systolic function with left ventricular ejection fraction estimated at 50%, mild mitral regurgitation; coronary arteries with normal origin and proximal dimensions, without evidence of hyperlucency, dilatation or aneurysms; and mild pericardial effusion.

In view of these findings, the diagnostic hypothesis of complete Kawasaki disease was raised and therapy with intravenous immunoglobulin, corticoid, antibiotics and acetylsalicylic acid was initiated. In this specific case, diuretic therapy with intravenous furosemide was implemented.

Furthermore, in this particular case, there was persistent elevation of inflammatory parameters despite timely treatment, so a single dose of Infliximab, a monoclonal antibody that inhibits TNF-alpha, was administered with subsequent improvement.

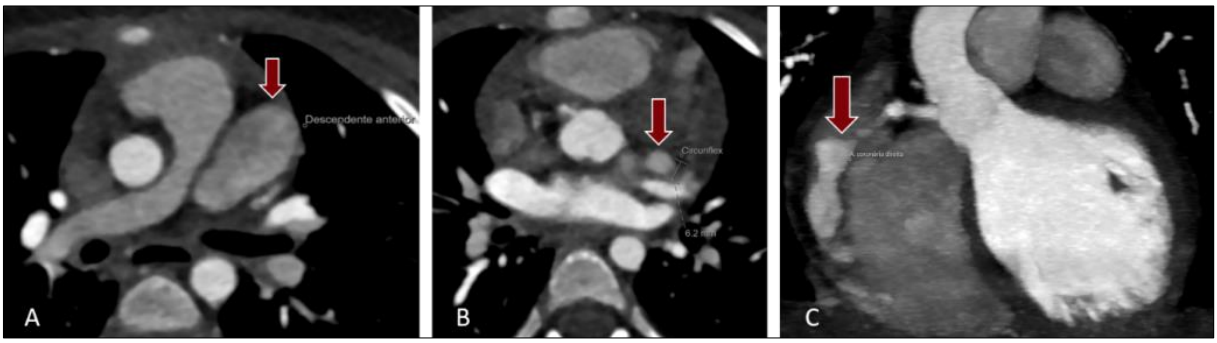
The patient maintained regular reassessment by pediatric cardiology and progressed to complete recovery of ventricular systolic function. However, the transthoracic echocardiogram performed on the 14th day of illness showed dilation of both coronary arteries, more significant on the left coronary artery. As seen in picture 1–A, the right coronary artery is dilated (3 mm, z-score +2.8) and there is a small aneurysm of the left coronary artery (4 mm, z-score +4.8).

The small aneurysm progressed to a giant aneurysm of the left coronary artery, with the maximum dilation verified on the 26th day of illness. In picture 1–B, the dilation of the right coronary artery remains similar, and there is a giant aneurysm of the left coronary artery (18 mm, z-score +41.3) and already showing areas of narrowing.



Picture 1: A) Transthoracic echocardiogram, parasternal short-axis view, showing diffuse dilation of right coronary artery (3 mm, z-score +2.8, yellow arrow) and small aneurysm of left coronary artery (4 mm, z-score +4.8, red arrow). B) Transthoracic echocardiogram, parasternal short-axis view, showing diffuse dilation of right coronary artery (3 mm, z-score +2.8, yellow arrow) and giant left coronary artery aneurysm (18 mm, z-score +41.3, red arrow) with areas of narrowing.

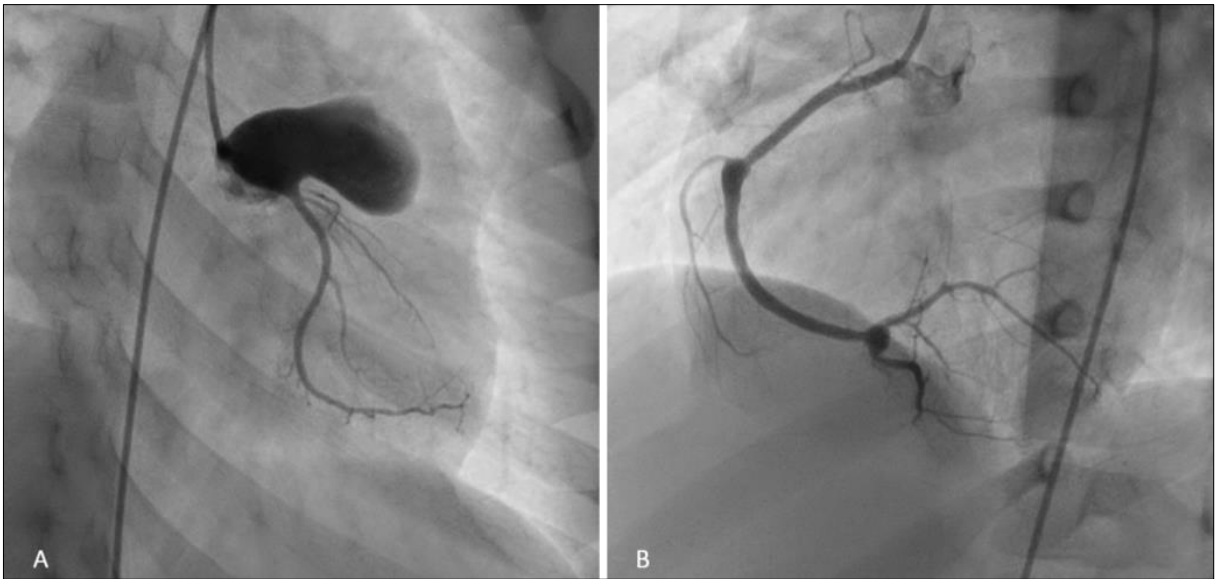
For a better anatomical characterization, cardiac CT angiography was performed on the 26th day of illness, showing a giant fusiform aneurysm of the left anterior descending artery (17 mm, z-score +43.2) – picture 2–A, focal dilation of the circumflex artery (6.2 mm, z-score +11.8) – picture 2–B, and diffuse fusiform aneurysms of the right coronary artery, the largest measuring 9 mm (z-score +18.0) – picture 2–C. It was noted a mild opacification of these aneurysms related to slow flow in the coronary circulation, however, without evidence of thrombosis.



Picture 2: A) Axial cardiac CT angiography image showing giant fusiform aneurysm of the left anterior descending artery (17 mm, z-score +43.2, red arrow). B) Axial cardiac CT angiography image showing focal dilation of the left circunflex artery (6.2 mm, z-score +11.8, red arrow). C) Coronal cardiac CT angiography image showing diffuse fusiform aneurysm of the right coronary artery (9 mm, z-score +18.0, red arrow)

Given these findings, he was medicated with hypocoagulation, antiplatelet aggregation, beta-blocker, besides control of cardiovascular risk factors. He remained asymptomatic and changes were never identified on the electrocardiogram, so he was discharged from the hospital, maintaining regular reassessment in consultation.

One year after the acute event, he underwent cardiac catheterization, which revealed a giant fusiform aneurysm of the left coronary artery (19 mm largest diameter, z-score +41.8), starting approximately at 5 mm from the ostium, involving the common trunk and extending to the anterior descending artery and proximal portion of the circumflex artery – picture 3–A; and two aneurysms of the right coronary artery, one in its middle portion and the other in its distal portion, measuring 3.7 mm (z-score +4.6) and 3.9 mm (z-score +5.7), respectively – picture 3–B.



Picture 3: A) Coronarography image showing giant fusiform aneurysm of the left coronary artery (19 mm largest diameter, z-score +41.8), involving the common trunk and extending to the anterior descending artery and proximal portion of the circumflex artery. B) Coronarography image showing two aneurysms of the right coronary artery, one in its middle portion (3.7 mm, z-score +4.6) and the other in it distal portion (3.9 mm, z-score +5.7)

Discussion

In this case, a previously healthy 3-year-old male presented with classical symptoms of Kawasaki disease, including prolonged fever, mucocutaneous manifestations such as conjunctivitis and changes in the oral mucosa, extremity changes, and cervical lymphadenopathy. The initial cardiac involvement consisted of depression of ventricular systolic function, and the severity of the disease manifested in the form of systemic involvement, with notable findings such as a multitude of laboratory abnormalities indicative of widespread inflammation and organ dysfunction.

Despite timely initiation of therapy with intravenous immunoglobulin, corticosteroids, antibiotics, and acetylsalicylic acid, the patient exhibited persistent elevation of inflammatory parameters, necessitating the administration of Infliximab, a monoclonal antibody targeting TNF-alpha. While this intervention led to improvement in the patient's condition, the subsequent development of coronary artery abnormalities underscores the importance of ongoing surveillance and aggressive management in Kawasaki disease cases with refractory or severe presentations.

The evolution of coronary artery involvement, from initial dilation to the progression of aneurysms, highlights the potential for long-term cardiac sequelae in Kawasaki disease survivors. The utilization of cardiac imaging modalities such as echocardiography and cardiac CT angiography provided crucial insights into the extent and morphology of coronary artery abnormalities, guiding subsequent therapeutic interventions aimed at reducing thrombotic risk and optimizing long-term outcomes.

Ultimately, this case underscores the need for heightened awareness among healthcare providers regarding the diverse clinical manifestations of Kawasaki disease and the importance of early recognition and aggressive management to mitigate the risk of serious complications, particularly coronary artery abnormalities. Additionally, it highlights the imperative for ongoing research efforts aimed at elucidating the pathogenesis of Kawasaki disease and identifying novel therapeutic strategies to improve outcomes in affected individuals.

Conclusion

The presented case report highlights the complex and potentially devastating consequences of Kawasaki disease. The possibility of progression to coronary aneurysms should always be considered even in the cases of adequate treatment and we should maintain regular imaging revaluation of these patients during the first weeks of disease.

Despite advancements in diagnosis and management, Kawasaki disease remains a significant clinical challenge, emphasizing the importance of ongoing research to elucidate its underlying pathogenesis and optimize therapeutic strategies for improved outcomes in affected children.

References

1. McCrindle BW, Rowley AH, Newburger JW, Burns JC, Bolger AF, Gewitz M, *et al.*; American Heart Association Rheumatic Fever, Endocarditis, and Kawasaki Disease Committee of the Council on

Cardiovascular Disease in the Young; Council on Cardiovascular and Stroke Nursing; Council on Cardiovascular Surgery and Anesthesia; and Council on Epidemiology and Prevention. Diagnosis, treatment, and long-term management of Kawasaki disease: a scientific statement for health professionals from the American Heart Association. Circulation. 2017 Apr 25;135(17).

2. Dallaire F, Dahdah N. New equations and a critical appraisal of coronary artery Z scores in healthy children. J Am Soc Echocardiogr. 2011 Jan;24(1):60-74.

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